

Introduction

Dermatomyositis (DM) and polymyositis (PM) are part of the idiopathic inflammatory myopathies, a heterogeneous group of chronic systemic autoimmune myopathies, associated with high morbidity and functional disability¹. In 1975, Bohan and Peter published a classic article that suggested a set of criteria to aid in the diagnosis and classification of DM and PM². Of the 5 criteria, 4 related to the muscle disease: (1) progressive proximal symmetrical weakness, (2) elevated muscle enzymes, (3) an abnormal **electromyography**, and (4) an abnormal muscle biopsy, while the fifth was the presence of compatible cutaneous disease. It was felt that DM differed from PM only by the presence of cutaneous disease². Recent studies of the myopathy suggests that the myopathies in DM and PM present different epidemiological, histological, immunohistochemical, pathological, and clinical characteristics, as well as different disease courses¹.

¹Shinjo SK.; Souza FHC.; Moraes JCB. Dermatomyositis and polymyositis: from immunopathology to immunotherapy (immunobiologics). Rv Bras Reumatol. 2013;53(1):103-110.
²Callen JP; Wortmann RL. Dermatomyositis. Clinics in Dermatology. 2006; 24:363-373

Clinical Case

53 years old female patient without relevant past clinical history

Attends to Emergency Department with a three month history of:

- ✓ generalized rash with pruritus:
 - predominantly in the trunk and upper limbs
 - extending to the face (close to the scalp), thighs and knees
- associated with:
 - lip swelling
 - asthenia
 - muscle fatigue
 - difficulty elevating the arms above shoulder level

Physical examination :

- V shaped rash in the upper torso;
- violaceous papular rash in the knees, lateral region of thighs, upper limbs, and nose;

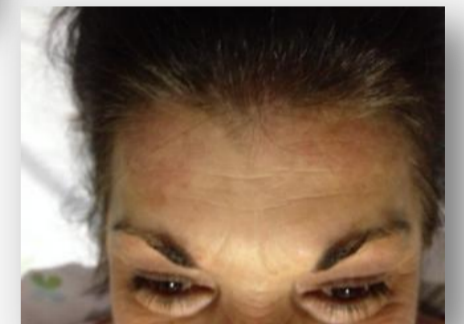
❖ **Blood analysis:** myoglobin of 312 ng/mL (without other abnormalities).

After admission:

- ✓ mild heliotrope
- ✓ muscle weakness of the pelvic and shoulder girdles

Initial study:

- ✓ increased muscle enzymes
- ✓ positive anti-nuclear antibodies (reactive 1/160 pattern).



Dermatomyositis ?

Muscle biopsy (triceps muscle) → atrophy of type II muscle fibres and perifascicular atrophy, necrosis, regeneration and hypertrophy of muscle cells

Skin biopsy → hyperkeratosis, acanthosis and epidermal atrophy, interface dermatitis, perivascular infiltrate of lymphocytes, dermal edema and deposit of mucin.

Electromyography → slight spontaneous activity (P waves and fibrillation) and in volunteer effort: motor units with myopathic features consistent with a framework of dermatomyositis

Echocardiogram → normal

Digestive endoscopy → Without alterations

Treatment:

- ✓ Prednisolone 1 mg/kg/day
- ↓
- rapid improvement of skin lesions and muscle symptoms;
 - normalization of the serum muscle enzyme
- Functional rehabilitation was started
 - Regular follow-up as an outpatient in the Autoimmune Diseases clinic

Conclusion

Dermatomyositis is a condition primarily of the skin and muscles, but other systemic features may occur. Dermatomyositis may also have inflammatory muscle diseases (polymyositis or inclusion body myositis)². The prognosis is good except for patients with associated malignancy, those with severe weakness and those with cardiac dysfunction, interstitial lung disease or the presence of a myositis-specific autoantibody other than Mi-2².

The correct interpretation of the clinical and analytical changes in an early stage enabled a fast diagnosis and a rapid initiation of therapy, minimizing the impact on the patients general health and quality of life.